

SURGERY FOR CONGENITAL HEART DISEASE

TRANSPPOSITION OF THE GREAT ARTERIES {S,D,L}

Pathologic anatomy, diagnosis, and surgical management of a newly recognized complex

The transposition of the great arteries {S,D,L} complex is delineated for the first time from the anatomic, diagnostic, and surgical standpoints in this study of 26 cases: 16 surgical and 10 postmortem. Transposition of the great arteries with situs solitus of the viscera and atria (S), D-loop ventricles (D), and L-transposition (L) was characterized by six additional interrelated anomalies that largely determined surgical management: (1) ventricular septal defect, usually conoventricular, in 96%; (2) malalignment of the conal septum, typically leftward and posteriorly, in 80%; (3) right ventricular hypoplasia in 50%; (4) pulmonary outflow tract stenosis in 27%; (5) ventricular malposition, such as superoinferior ventricles, in 23%; and (6) absent left coronary ostium resulting in "single" right coronary artery in 23%. Complete surgical repair was done in 81% of the surgical patients with a 12.5% hospital mortality rate and no late deaths. When there was no pulmonary outflow tract stenosis and intracardiac anatomy was uncomplicated, we undertook anatomic repair before 1 month of age. However, when pulmonary outflow tract stenosis coexisted, complete repair was deferred until after age 1 year, our currently preferred operation being the REV procedure (*réparation à l'étage ventriculaire*). When complex intracardiac anatomy precluded biventricular repair, a palliative procedure was performed in 19% without mortality. Hence, this experience indicates that surgical management of patients with the transposition of the great arteries {S,D,L} complex is feasible. (*J THORAC CARDIOVASC SURG* 1995;110:613-24)

Lucile Houyel, MD,^a Richard Van Praagh, MD,^b
Francois Lacour-Gayet, MD,^a Alain Serraf, MD,^a Jérôme Petit, MD,^a
Jacqueline Bruniaux, MD,^a and Claude Planché, MD,^a *Paris, France, and Boston, Mass.*

In the commonest form of physiologically uncorrected (complete) transposition of the great arteries (TGA), the segmental set consists of viscerotransposition (S), D-loop ventricles (D), and D-transposition (D), that is, TGA {S,D,D}. The segmental alignments typically are atrioventricular concordance and ventriculoarterial discordance.

Infrequently, however, the transposed aortic valve can lie to the left of the transposed pulmonary valve, resulting in TGA {S,D,L}. The presence of L-TGA may initially suggest that congenitally physiologically corrected TGA is present. However, TGA {S,D,L} typically also has atrioventricular concordance and ventriculoarterial discordance. Because there is only one segmental alignment discordance (rather than two), this fact indicates that TGA {S,D,L} is congenitally physiologically uncorrected (rather than congenitally physiologically corrected).

Nonetheless, physiologically uncorrected L-TGA in viscerotransposition is sufficiently rare to be potentially confusing both diagnostically and surgically. TGA {S,D,L} with two patent atrioventricular valves and two ventricular sinuses accounted for only 2.9% of the cases of physiologically uncor-

From Hôpital Marie-Lannelongue, Paris, France,^a and Children's Hospital and Harvard Medical School, Boston, Mass.^b

Received for publication Oct. 12, 1994.

Accepted for publication Jan. 18, 1995.

Address for reprints: Lucile Houyel, MD, Hôpital Marie-Lannelongue, 133 avenue de la Résistance, 92350 Le Plessis-Robinson, France.

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0022-5223/95 \$5.00 + 0 12/1/63553

Table I. Clinical data of 16 surgical patients with TGA {S,D,L}

Case No.	Sex	Age at operation	Previous palliative operation	Type of operation	Follow-up data
1	M	7 yr	BT shunt, surgical ASD (1 yr)	Rastelli (1981)	RV-PA conduit replacement (1992); doing well
2	F	6 mo	MPA band (1-2 mo)	ASO (1984)	Doing well
3	F	1.2 mo		ASO (1984)	Postop. death
4	F	6 yr	Left BT shunt	Right MBT shunt (1984)	Doing well
5	F	10 mo		ASO (1984)	Intraop. death
6	F	4 days		Senning (1984)	Doing well
7	F	2 mo		ASO (1984)	Doing well
8	F	19 days		ASO (1985)	Doing well
9	M	10 mo	MPA band (3 mo)	ASO (1986)	Doing well
10	F	20 days		ASO (1990)	Doing well
11	M	16 mo	MPA band, CoAo repair, PDA lig. (5 days)	Bid. Glenn (1991)	MPA division (6 mo postop.); doing well
12	M	10 mo	MPA band, CoAo repair (18 days)	ASO (1991)	MPA band (14 days postop.); VSD closure (1994); doing well
13	M	7 mo		ASO (1992)	Doing well
14	M	7 days		ASO (1993)	Doing well
15	F	7 mo	MPA band, PDA lig. (4 days)	Right MBT shunt (1993)	Doing well
16	M	4 yr	Right MBT shunt (9 mo)	REV (1994)	Doing well

ASD, Atrial septal defect; ASO, arterial switch operation; Bid., bidirectional; BT, Blalock-Taussig; CoAo, coarctation of the aorta; F, female; lig., ligation; M, male; MBT, modified Blalock-Taussig; MPA, main pulmonary artery; PA, pulmonary artery; PDA, patent ductus arteriosus; RV, right ventricle.

rected TGA with viscerotrial situs solitus recorded in the Cardiac Registry of the Children's Hospital in Boston, Massachusetts (10 of 347 cases).

This study is focused on TGA {S,D,L} because of the relevance of an understanding of the pathologic anatomy to successful surgical repair by means of the arterial switch, the Rastelli, or the REV (réparation à l'étage ventriculaire) procedures.

Patients and methods

This study was based on 26 cases of TGA {S,D,L}: 16 in patients operated on at Hôpital Marie-Lannelongue, Le Plessis-Robinson, France, and 10 postmortem cases from the Cardiac Registry, Children's Hospital, Boston, Massachusetts. Only cases with two patent atrioventricular valves and two ventricles were included in this surgically oriented study.

The operations were performed between 1981 and 1994, inclusive, by the same surgical team. Age at operation ranged from 4 days to 7 years, with a median of 8 months and a mean of 18 months.

During study of the pathologic anatomy of the 10 postmortem heart specimens, particular attention was paid to the cardiac geometry^{1,2} as follows: (1) the ventricular septal angle^{3,4} is the angle between the planes of the ventricular and the atrial septa, (2) the conoventricular septal angle⁵ is the angle between the conal septum (infundibular septum or parietal band) and right ventricular sinus septal surface, and (3) the rotation of the aortic valve^{1,2,5} was measured. A needle was passed through the aortic wall immediately above the middle of the noncoronary leaflet and then advanced through the intercoronary

commissure at the aortopulmonary septum. The angle of this diameter of the aortic valve relative to the sagittal plane was then measured with a circular protractor in four-chamber long-axis projection.

The coronary ostia were often helpful in measuring the rotation of the aortic valve. When both coronary ostia were present, the intercoronary line, a line between the coronary ostia, was readily demarcated. The angle between the intercoronary line and the frontal plane indicated the levorotation of the aortic valve (in degrees).

Statistical analysis was done with the χ^2 test with the Yates correction and, when appropriate, Fisher's exact test was used. A *p* value <0.05 was regarded as statistically significant.

Results

The findings are presented in detail in the tables (Tables I through IV), are illustrated in the figures (Figs. 1 through 7), and are summarized as follows.

The heart was left-sided (levocardia) in 24 (92.3%) of 26 cases (Fig. 1) and right-sided (dextrocardia) in 2 cases (7.7%). A secundum type of atrial septal defect occurred in 11 (42.3%) of 26 cases and a surgically or balloon-created atrial septal defect in the remaining 15 (57.7%) of 26 cases. Left juxtaposition of the atrial appendages was present in 4 (15.4%) of 26 cases (Table III, Fig. 1). The left ventricle was always well developed (Fig. 2, B, Fig. 3, B, and Fig. 7). The right ventricle was hypoplastic in 50% of patients (13 of 26 cases, Table III, Fig. 6). Superoinferior ventricles were present in 19.2% (5

of 26 cases, Table III, Fig. 6). Crisscross atrioventricular relations occurred in 1 patient (3.8%, Table III).

Ventricular malposition. Malposition of the ventricles relative to the atria was most marked with superoinferior ventricles (Fig. 6) and with crisscross atrioventricular relations. The ventricular malposition with superoinferior ventricles was quantitated in the postmortem cases by means of the ventriculoatrial septal angle (Table IV): case 19, 115 degrees to the left; case 21, 100 degrees to the left; and case 21, 100 degrees to the left. These very large angles quantitated the marked malposition of the ventricles relative to the atria.

Ventricular septal defect. Ventricular septal defect (VSD) was present in 25 (96.2%) of the 26 cases; the only exception was case 8 (Tables I and III). The VSD was almost always conoventricular in type, that is, occurring between the conal septum above and the ventricular septum and septal band below (24 [96%] of 25 cases, Figs. 2 and 3). The VSD was never purely membranous, that is, a deficiency of the membranous septum only. One patient had a small midmuscular VSD only (3.8%). Multiple VSDs occurred in two patients (7.7%). A confluent conoventricular plus atrioventricular canal type of VSD was found in three patients (11.5%, Table III). Each had a small right ventricular sinus and a straddling tricuspid valve.

The aortic valve overrode a conoventricular type of VSD in two patients (Table III). These two cases were reminiscent of double-outlet left ventricle of the {S,D,L} type,⁶ except that the overriding aorta originated predominantly above the right ventricle.

Conus. A subaortic muscular conus (Fig. 3, A) was present in 25 (96.2%) of the 26 cases; the only exception was one patient (case 5, Table III) who had a bilateral conus (subaortic and subpulmonary). This patient also had left juxtaposition of the atrial appendages.

All patients with a VSD also had conal septal malalignment relative to the underlying ventricular septum and septal band (Figs. 2 and 3). Conal septal malalignment consisted of abnormal conal septal angulation or abnormal conal septal displacement, or both (Table IV), and both usually occurred in a leftward and posterior direction (Fig. 2, B, and Fig. 3, B). In the eight postmortem cases in which this could be measured, the mean conoventricular septal angle ± 1 standard deviation was 71 ± 44 degrees (Table IV); normal is 56 ± 26 degrees. Hence only three of these conoventricular septal angles were outside the normal range: 160 degrees left in case

Table II. Clinical data in 10 postmortem cases of TGA {S,D,L}

Case No.	Sex	Age at death	Previous surgical procedures (age)
17	M	10 mo	None
18	M	2.5 mo	None
19	M	1.5 mo	MPA band, surgical ASD (1.5 mo)
20	M	9 mo	MPA band (1 mo)
21	M	4 mo	MPA band, CoAo repair, PDA lig. (5 days)
22	M	24 yr	Left BT shunt (5.5 yr) Right BT shunt (13 yr)
23	F	12 mo	PDA lig. (17 days) MPA band (19 days) Senning (1 mo)
24	M	21 yr	MPA band (14 days) Right BT shunt (6.5 yr) Mustard (9.5 yr)
25	M	17 yr	Blalock-Hanlon, Right BT shunt (19 mo) Mustard (7.5 yr) TV replacement (17 yr)
26	NK	NK	Right BT shunt, surgical ASD, Ao-LPA anastomosis

Ao, Aorta; ASD, atrial septal defect; BT, Blalock-Taussig; CoAo, coarctation of aorta; F, female; lig., ligation; LPA, left pulmonary artery; M, male; MPA, main pulmonary artery; NK, not known; PDA, patent ductus arteriosus; TV, tricuspid valve.

17, 0 degrees in case 18, and 90 degrees left in case 26 (Table IV). However, in the nine postmortem cases in which it could be observed, conal septal displacement to the left was present in all, and posterior displacement of the conal septum was found in 7 (78%) of the 9 cases (Table IV). Conal septal hypoplasia was present in 12 (46%) of the 26 cases (Table III).

Subpulmonary stenosis occurred in 7 (27%) of the 26 cases. Subpulmonary stenosis resulted from posterior displacement of the conal septum in all (Fig. 5) and was associated with subvalvular fibrous tissue in one (case 26, Table III, Fig. 2, B). Two patients also had pulmonary valvular stenosis with a bicuspid (bicommissural) pulmonary valve.

Coronary arteries. Because of the levorotation of the L-transposed aortic valve, the right coronary ostium was anterior to the left coronary ostium, as in typical TGA {S,L,L}.

The levorotation of the L-transposed aortic valve relative to the sagittal plane varied from 20 to 75 degrees to the left, with a mean of 40 ± 19 degrees and a median of 41 degrees (Table IV) (Fig. 4).

The distributions of the coronary arteries corresponded to those of D-loop ventricles, with the

Table III. Anatomic findings in 26 cases of TGA {S,D,L}

Case No.*	Location of VSD	Hypoplasia of conal septum	Conal septal malalignment	Subpulmonary stenosis	Right ventricle hypoplasia	Superoinferior ventricles	Coronary arteries	Other associated anomalies
1	CV-SubAo	+	+	+	—	—	U	Overriding Ao
2	CV-SubAo	—	+	—	—	—	U	LJAA
3	CV-SubAo	—	+	—	+	—	U	
4	CV-SubAo	—	++	+	+	+	U	Dextrocardia
5	CV-SubAo	Bilat. conus	+	—	—	—	U	LJAA
6	CV-NonC	—	—	—	—	—	U	
7	CV-SubAo	+	+	—	—	—	U	RPA stenosis
8		—	—	—	—	—	U	
9	CV-SubP	—	+	—	—	—	U	Ao arch hypoplasia
10	Midmuscular	—	—	—	—	—	U	
11	CV-Doubly committed	+	—	—	++	—	U	CoAo
12	CV-AVC-Midmuscular	—	—	—	+	+	Left circumflex— from RCA	Straddling TV-CoA
13	CV	+	+	—	—	—	U	LJAA
14	CV	—	—	—	—	—	"Single" RCA	
15	CV-AVC	+	+	—	+	+	"Single" RCA	Overriding TV
16	CV-Midmuscular	+	+	+	—	—	U	
17	CV-SubAo	++	+	+	++	—	"Single" RCA	Overriding Ao
18	CV-SubP	—	+	—	++	—	U	LJAA
19	CV-SubAo	+	+	—	++	+	U	
20	CV-SubP	—	+	—	+	—	"Single" RCA	
21	CV-SubP	—	+	—	++	+	U	CoAo
22	CV-SubAo	++	++	++	++	—	"Single" RCA	Dextrocardia plus straddling MV, crisscross
23	CV-SubAo	+	+	—	+	—	U	
24	CV-SubP	—	+	—	—	—	U	
25	CV-SubAo-AVC	+	NK	+	+	—	"Single" RCA	Straddling TV
26	CV-SubAo	++	+	+	—	—	U	

Ao, Aorta; AVC, atrioventricular type; *bilat.*, bilateral; *CoAo*, coarctation of aorta; *CV*, conoventricular; *LJAA*, left juxtaposition of the atrial appendages; *MV*, mitral valve; *NK*, not known; *NonC*, noncommitted; *RCA*, right coronary artery; *RPA*, right pulmonary artery; *SubAo*, subaortic; *subP*, subpulmonary; *TV*, tricuspid valve; *U*, usual; +, mild to moderate; ++, marked.

*Cases 1 to 16, inclusive, are living postoperative patients; cases 17 to 26, inclusive, are postmortem cases.

Table IV. Geometric measurements in 10 postmortem cases of TGA {S,D,L}

Case No.	Ventriculoatrial septal angle in degrees (normal ≤ 50 degrees)	Conoventricular septal angle in degrees (normal 56 ± 26 degrees)	Conal septal displacement (normal 0)	Aortic valve rotation in degrees (normal 150 degrees R)
17	10 L	160 L	L & post.	75 L
18	0	0	L & post.	20 L
19	115 L	70 L	L & post.	50 L
20	50 L	55 L	L	23 L
21	100 L	60 L	L	35 L
22	75 L	70 L	L & post.	50 L
23	NM	60 L	L & post.	20 L
24	NM	NM	L & post.	47 L
25	NM	NM	NM	20 L
26	45 L	90 L	L & post.	60 L

L, left; NM, not measured because of surgical artifact; *post.*, posterior; R, right.

right coronary artery supplying the right ventricle and the left coronary artery supplying the left in the majority of cases (19 [73%] of 26). This we called the usual coronary artery distribution (Table III).

Coronary anomalies were found in the remaining 7 (27%) of the 26 cases. The most frequent anomaly was absence of the left coronary ostium (so-called single right coronary artery), which was observed in 6 of these 7 cases (Table III). In the seventh patient (case 12, Table III), the circumflex artery originated from the right coronary ostium along with the right coronary artery. The circumflex artery then ran posteriorly and to the left, passing behind the transposed pulmonary artery to reach the left atrioventricular groove posteriorly.

Two cases had a straddling tricuspid valve (7.7%): cases 12 and 25 (Table III). The VSD was the

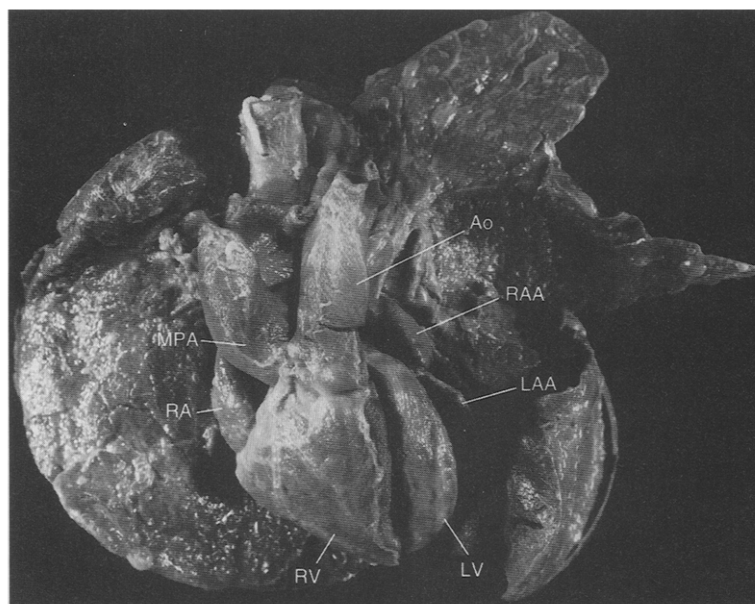


Fig. 1. Exterior of heart specimen of case 18 (Tables II and III), 2.5-month-old boy, seen from front. Morphologically right atrium (*RA*) is right-sided, but right atrial appendage (*RAA*) and left atrial appendage (*LAA*) are left-sided, indicating viscerotransposition with left juxtaposition of atrial appendages. Morphologically right ventricle (*RV*) is right-sided and morphologically left ventricle (*LV*) is left-sided, denoting D-loop ventricles. Aorta (*Ao*) arises from right ventricle but is anterior and to left relative to large main pulmonary artery (*MPA*), which originates from left ventricle. Hence external inspection suggests correct diagnosis: TGA {S,D,L}, with left juxtaposition of atrial appendages and without pulmonary stenosis.

confluent conoventricular plus atrioventricular canal type in both. One patient (case 15, Table III) had an overriding tricuspid valve with chordal insertions into the crest of the ventricular septum. One case (case 22, Table III) had a mitral valve that straddled through a conoventricular type of VSD. All of the aforementioned four cases with a straddling or overriding atrioventricular valve also had a small right ventricular sinus (Table III).

Coarctation of the aorta was present in 3 (11.5%) of the 26 cases (Table III). Although obvious aortic infundibular or valvular stenosis was not present in any of these cases, all had small right ventricles, which may be regarded as a form of subaortic stenosis.

Hypoplasia of the aortic arch was found in one patient (case 9, Table III). The VSD was subpulmonary, no pulmonary outflow tract stenosis was present, and blood appeared to have flowed preferentially into the pulmonary artery.

Surgical series. Among the 16 surgical patients, the diagnosis of TGA {S,D,L} was established preoperatively in the majority (10 [62.5%] of 16). One palliative procedure was done in 8 (50%) of these 16

patients before complete repair: pulmonary artery banding in 5 and a modified Blalock-Taussig shunt in 3 (Table I). In 13 (81%) of these 16 patients, complete repair was carried out (Table I).

The major problem encountered during the arterial switch operation (10 patients) had to do with closure of the VSD, which could not be done through the right atrium in five patients; thus ventriculotomy was required in three (cases 3, 5, and 13) and a transpulmonary approach in two (cases 9 and 12, Tables I and III).

The Senning procedure was performed early in our experience (1984) in one patient (Table I) because the main pulmonary trunk was judged too short to permit an arterial switch with the Lecompte maneuver (anterior translocation of the pulmonary trunk).

One patient with VSD and subpulmonary stenosis underwent a Rastelli procedure (1981). A REV procedure was done in another patient with VSD and subpulmonary stenosis (case 16, Tables I and III) and consisted of the following: (1) enlargement of the VSD by resection of the conal septum, (2) construction of an intraventricular tunnel from the

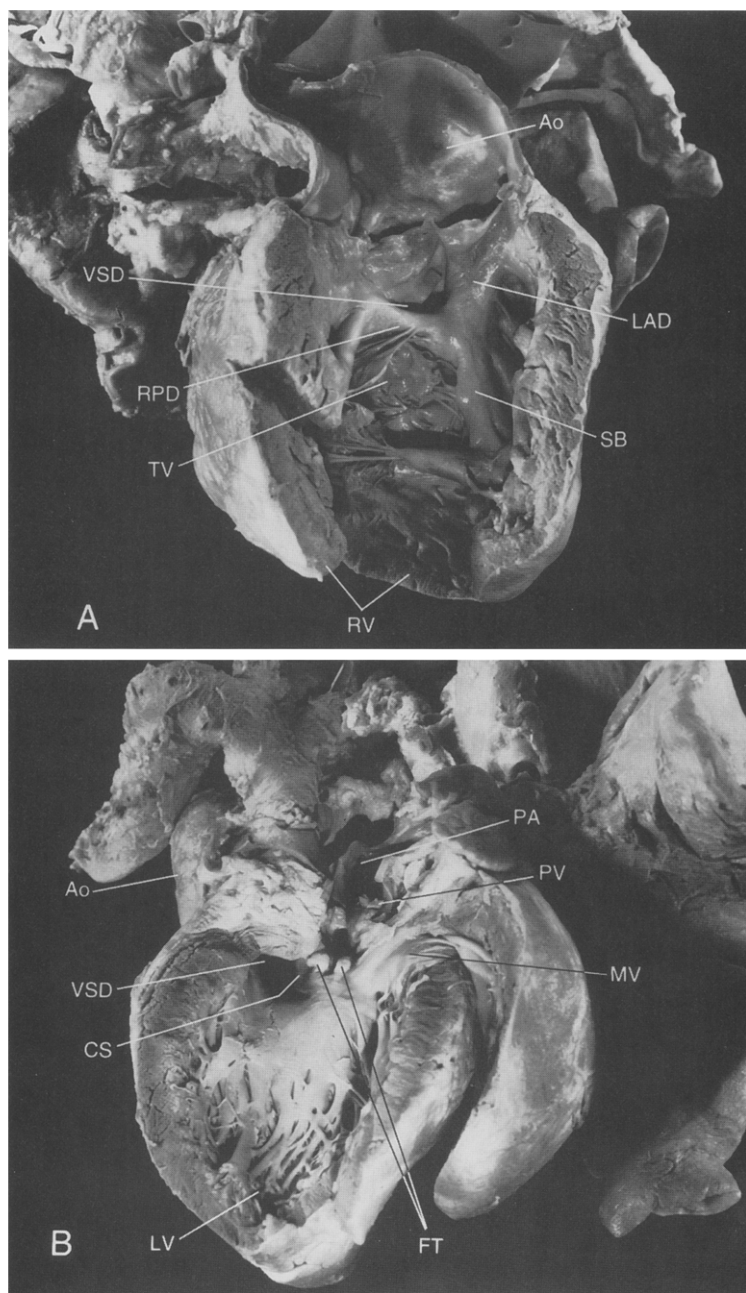


Fig. 2. Heart specimen of case 26 (Tables II and III). **A**, Opened right ventricle (*RV*) and L-transposed aorta (*Ao*). Note that VSD is subaortic and that its inferior margins are formed by prominent right posterior division (*RPD*) and left anterior division (*LAD*) of septal band (*SB*). VSD is not paramembranous, that is, not confluent with tricuspid valve (*TV*) because of prominent right posterior division of septal band. Subsemilunar conal septum (parietal band) is not well seen. **B**, Opened left ventricle (*LV*), stenotic pulmonary outflow tract, and hypoplastic pulmonary artery (*PA*). Conal septum (*CS*) is markedly hypoplastic and malaligned leftward and posteriorly, "squeezing" subpulmonary outflow tract. Conal septum forms right angle relative to underlying ventricular septum (Table IV), with conal septum running into subpulmonary outflow tract. VSD is subaortic because of leftward and posterior malalignment of hypoplastic conal septum. Where conal septum abuts pulmonary outflow tract, fibrous tissue (*FT*) contributes to subpulmonary stenosis. Bicuspid pulmonary valve (*PV*) is in direct fibrous continuity with anterior leaflet of mitral valve (*MV*).

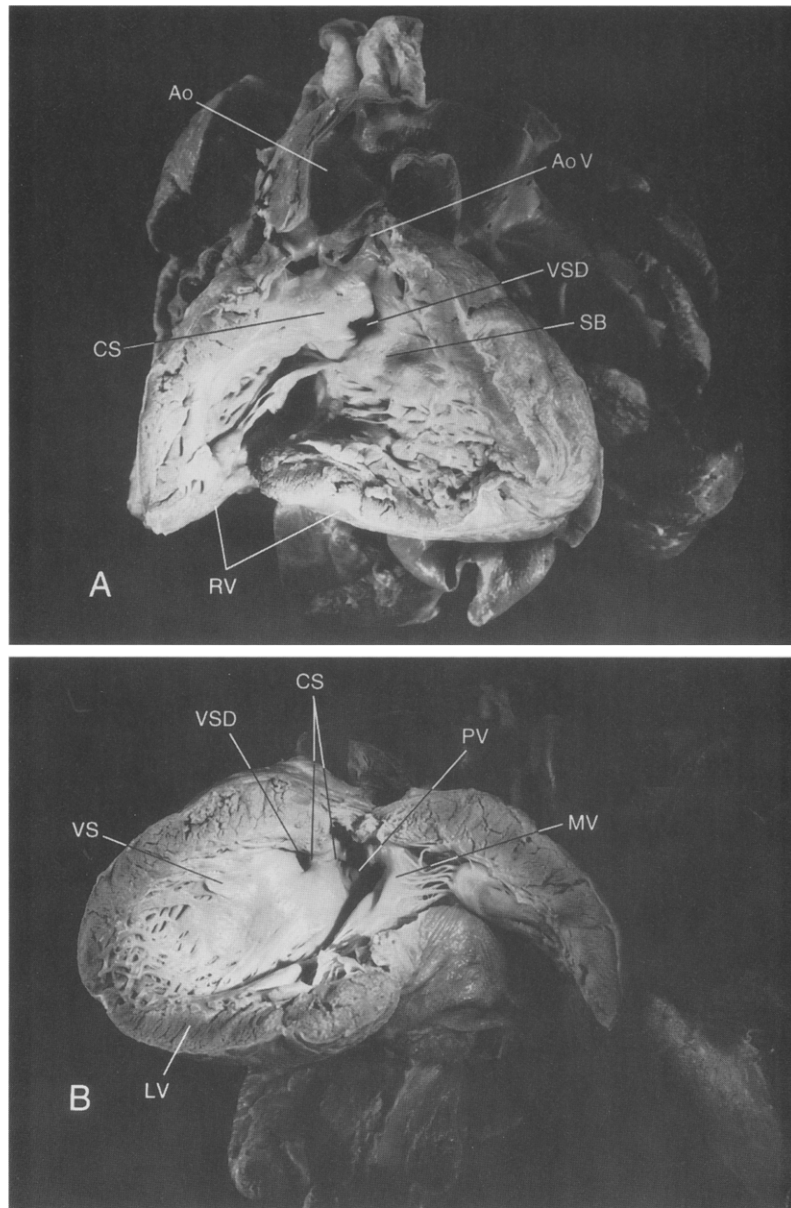


Fig. 3. **A**, Opened right ventricle (*RV*) and L-transposed aorta (*Ao*) of case 20, 1.5-month-old boy (Tables II through IV). Conal septum (*CS*) is well developed, is not malaligned far to left, and hence is readily visible from right ventricular perspective. VSD is conoventricular in type, between conal septum above and ventricular septum and septal band (*SB*) below. Aortic valve (*AoV*) is widely separated from VSD by subaortic conus. **B**, Opened left ventricle (*LV*), showing inferior surface of conal septum (*CS*) that is angulated 55 degrees leftward relative to ventricular septum (*VS*) (Table IV) and that is displaced only mildly posteriorly (Table IV). Right surface of the conal septum is flush with left surface of ventricular septum. There is direct fibrous continuity between pulmonary valve (*PV*) and mitral valve (*MV*). VSD is closer to pulmonary valve than to aortic valve (in **A**), but VSD is not truly subpulmonary because of leftward and posterior malalignment of conal septum; hence VSD may be described as “subpulmonary.” Subpulmonary left ventricular outflow tract is widely patent because conal septal malalignment is mild.

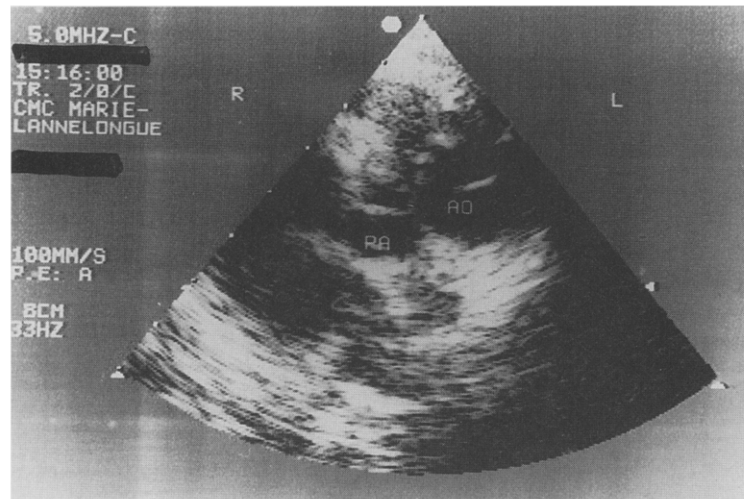


Fig. 4. Two-dimensional echocardiographic study of case 15, 7-month-old girl (Tables I and III). Parasternal short-axis view shows anterior and leftward aorta (*Ao*) and posterior and rightward pulmonary artery (*PA*), typical of L-TGA.

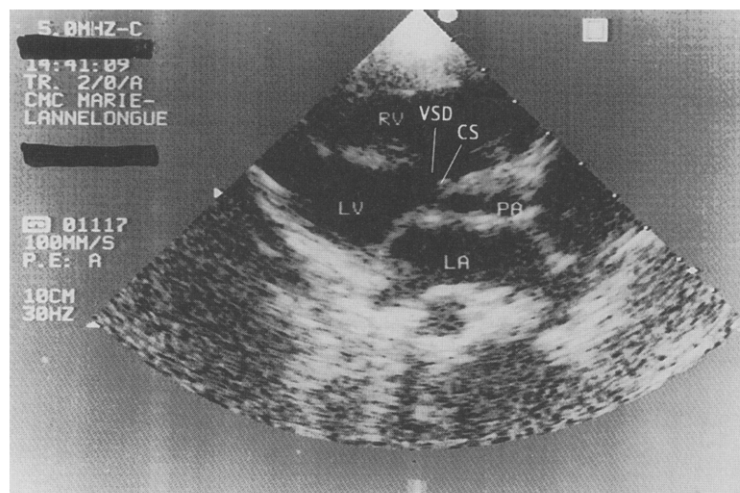


Fig. 5. Two-dimensional echocardiographic study of case 16, 4-year-old boy (Tables I and III). Parasternal long-axis view shows large subaortic VSD and posterior malalignment of conal septum (*CS*) with subpulmonary stenosis. *LA*, Left atrium; *LV*, left ventricle; *PA*, pulmonary artery; *RV*, right ventricle.

left ventricle to the transposed aortic valve by means of a prosthetic patch, and (3) a Lecompte maneuver followed by direct implantation of the pulmonary trunk into the right ventricle.

In three patients (19%), only palliative procedures were done because of the complexity of the intracardiac lesions (cases 4, 11, and 15, Tables I and III).

Two of these 16 surgical patients died after operation, both after the arterial switch procedure, giving a hospital mortality rate of 12.5%. One patient,

a 10-month-old girl (case 5, Tables I and III), had severe pulmonary hypertension before operation. However, because a lung biopsy specimen was interpreted as showing negative results, complete repair was undertaken. The cause of death in the other patient, a 1.2-month-old girl (case 3, Tables I and III), remains unknown. There were no late deaths.

All 14 surviving patients were found to be doing well at postoperative follow-up (Table I). Follow-up examination included two-dimensional echocardiog-

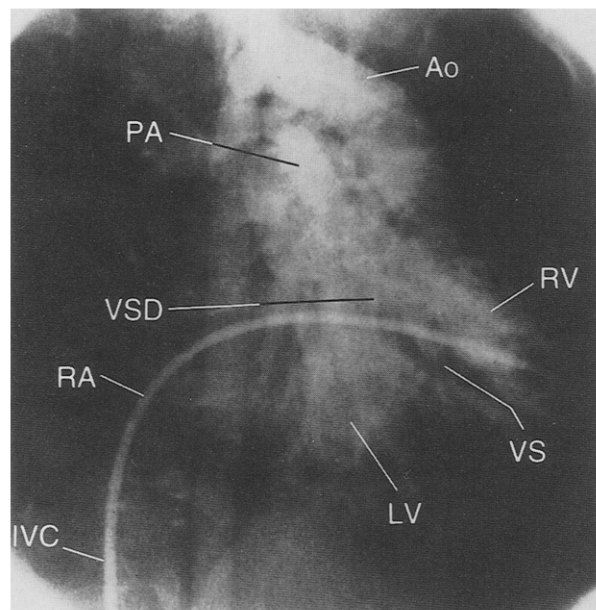


Fig. 6. Selective right ventricular (RV) angiogram in posteroanterior projection in case 12, 10-month-old boy (Tables I and III). Right ventricle is superior and small, left ventricle (LV) is inferior and large, and ventricular septum (VS) is approximately horizontal, indicating that ventricular malposition with superoinferior ventricles is present. Inferior vena cava (IVC) and right atrium (RA) are right-sided, indicating situs solitus of viscera and atria. Catheter passed from right atrium to small and superior right ventricle, suggesting concordant atrioventricular alignments. Aorta (Ao) arises from right ventricle to left of large and unobstructed pulmonary artery (PA) that originates from left ventricle. VSD is large and subpulmonary. (See Table III, case 12, for additional anatomic findings.)

raphy in all patients and cardiac catheterization in half (7 patients).

In three patients (21.4%) reoperation was required (detailed in Table III), and there was no operative mortality. Replacement of a right ventricular–pulmonary artery conduit was done in case 1 (Tables I and III) 11 years after the Rastelli procedure had been done. Division of the main pulmonary artery was done in case 11 (Tables I and III) 6 months after a bidirectional Glenn procedure was done because of the persistence of significant flow through the ligated main pulmonary artery. In case 12 (Tables I and III), pulmonary artery banding was necessary 14 days after an arterial switch operation because of multiple residual VSDs. Three years later this patient underwent successful closure of the VSDs and main pulmonary artery band removal.

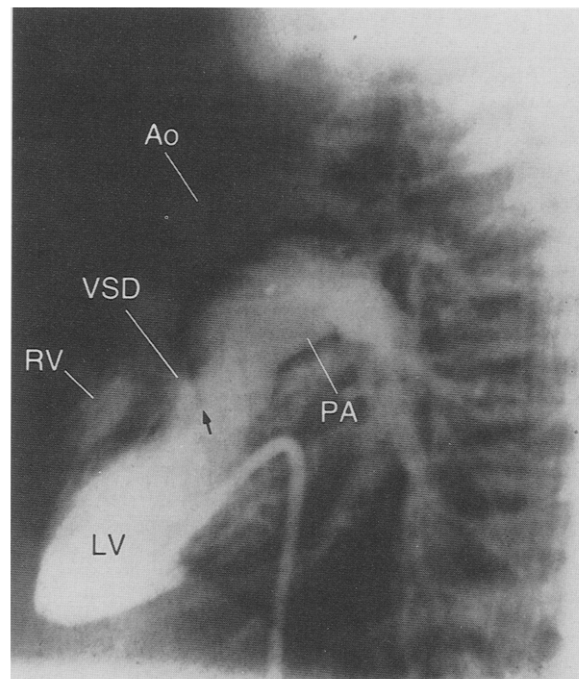


Fig. 7. Selective left ventricular (LV) angiogram, left lateral projection, in case 9, 10-month-old boy (Tables I and III). Transposed pulmonary artery (PA) arises from well-developed left ventricle. Conal septum is mildly posteriorly displaced (arrow), without producing subpulmonary stenosis. Also seen are conoventricular type of VSD that is subpulmonary, right ventricle (RV), and transposed aorta (Ao).

Discussion

TGA {S,D,L} was often associated with six inter-related associated anomalies (Table V): (1) VSD, typically conoventricular, in 96% (Figs. 2, 3, and 5); (2) conal septal malalignment, usually leftward and posteriorly, in 80% (Fig. 2, B, Fig. 3, B, and Fig. 6); (3) right ventricular hypoplasia in 50% (Fig. 6); (4) pulmonary outflow tract stenosis in 26.9% (Fig. 2, B, and Fig. 5); (5) ventricular malposition, as with superoinferior ventricles, in 23% (Fig. 6); and (6) absent left coronary ostium, resulting in “single” right coronary artery, in 23%.

Each of these characteristic additional malformations was associated significantly more frequently with TGA {S,D,L} than with the classic form of physiologically uncorrected transposition, that is, TGA {S,D,D} (Table V, $p < 0.01$ to 0.02). As will be seen, these anomalies (Table V) largely determined the appropriate surgical management of the cases.

Left juxtaposition of the atrial appendages^{11,12} (Fig. 1) was present in 15.4% of patients with TGA

Table V. Comparison of TGA {S,D,L} and TGA {S,D,D}

Anatomic finding	TGA {S,D,L} (%)	TGA {S,D,D} (%)*	p Value
1. VSD	96	14.8 ⁷ -31 ⁸	<0.01
2. Conal septal malalignment	80	21 ⁸	<0.01
3. Right ventricular hypoplasia	50	0 ⁷⁻⁹	<0.01
4. Pulmonary outflow tract stenosis	27	9.7 ⁹	<0.01
5. Ventricular malposition: superoinferior ventricles and crisscross AV relations	23	0 ⁷⁻⁹	<0.01
6. Absent left coronary arterial ostium ("single" RCA)	23	4.5 ¹⁰ -4.8 ⁸	<0.02

AV, Atrioventricular; RCA, right coronary artery; VSD, ventricular septal defect.

*Prevalences in TGA {S,D,D} from the literature, indicated by references.

{S,D,L} (Table III), but this was not statistically significantly different from its prevalence with TGA {S,D,D} (4.8%,⁹ $p = 0.9$).

From the anatomic and developmental standpoints, TGA {S,D,L} appears to be not only a malformation of the conus (infundibulum),^{13, 14} but also often a malformation of the ventricular part of the heart.¹⁵ Involvement of the ventricular segment is indicated by the high prevalences of right ventricular hypoplasia (50%, Table V) and ventricular malposition (23%, Table V). The high prevalences of VSD (mainly at the conoventricular junction, 96%, Table V), conal septal malalignment (usually leftward and posteriorly, 80%, Table V), and pulmonary outflow tract stenosis (27%, Table V, related to leftward and posterior conal septal malalignment) all then become understandable.

Absence of the ostium of the left coronary artery, resulting in "single" right coronary artery (23%, Table V), may be related developmentally to the disadvantageous location of the left coronary arterial bud from the left coronary sinus of Valsalva in association with L-TGA. Levorotation of the aortic valve places the left coronary bud posteriorly, adjacent to the anterior atrial wall: behind the left anterior atrioventricular sulcus, where the coronary angioblasts are located with which the left coronary bud usually anastomoses to form the left circumflex coronary artery. Our hypothesis therefore is that the abnormally leftward and posterior location of the left coronary bud, adjacent to the atrial wall, predis-

poses to nonconnection between the left coronary bud and the coronary angioblasts in the anterior left atrioventricular sulcus. Hence the left coronary bud involutes and the anterior right-sided coronary bud develops and anastomoses with all of the coronary angioblasts in the interventricular and atrioventricular sulci. Consequently, TGA {S,D,L} is associated with an unusually high prevalence of absent left coronary ostium and its sequela: a "single" right coronary artery (23%, Table V).

L-transposition of the great arteries in association with D-loop ventricles, as in TGA {S,D,L}, is a "flag" that indicates that ventricular malposition may well be present. L-TGA with D-loop ventricles is an exception to the loop rule.^{1, 2} This rule summarizes the usual correlations between the great arteries and the ventricles: (1) when the aortic valve is right-sided relative to the pulmonary valve, D-loop ventricles usually are present and (2) when the aortic valve is left-sided relative to the pulmonary valve, L-loop ventricles usually are present.

Any exception to the loop rule is unusual, by definition, and often interesting. For example, in superoinferior ventricles,¹⁵ we found that exceptions to the loop rule are frequent (70%); exceptions occurred in three cases of TGA {S,D,L}. With superoinferior ventricles, exceptions to the loop rule became the rule.¹⁶ The question remained: why? These exceptional aortopulmonary relationships were unusual because the ventricles, from which the great arteries originated, were themselves abnormally located in space.

It must be added, however, that there are many different kinds of exceptions to the usual arterioventricular relationships (summarized by the loop rule), not all of which are associated with ventricular malpositions.^{5, 17}

The main difference between TGA {S,D,L} and typical TGA {S,D,D} is that the ventricular segment is often abnormal in TGA {S,D,L} (Table V), whereas this is seldom the case in classic TGA {S,D,D}.

L-TGA in association with D-loop ventricles plus the characteristic six related additional anomalies (Table V) may be regarded as a complex¹⁸: the TGA {S,D,L} complex.

The degree of levorotation of the infundibulum and great arteries relative to the underlying ventricles and ventricular septum has several important consequences that are illustrated by the differences between case 26 (Fig. 2) and case 20 (Fig. 3). The degree of conal septal levorotation, posterior dis-

placement, and hypoplasia was much greater in case 26 (Fig. 2) than in case 20 (Tables III and IV). The levorotation of the transposed aortic valve was also much greater in case 26 than in case 20 (Table IV). The ventricular septal defect was subaortic in case 26 with greater infundibuloarterial levorotation and was closer to the pulmonary valve (although not truly subpulmonary) in case 20 with less infundibuloarterial levorotation. In addition, the caliber of the pulmonary outflow tract was stenotic in case 26 with greater infundibuloarterial levorotation and was widely patent in case 20 with less infundibuloarterial levorotation.

Hence with TGA {S,D,L} it was found that infundibuloarterial levorotation varied directly with (1) subaortic VSD location (as opposed to "subpulmonary" location), (2) pulmonary outflow tract stenosis (as opposed to wide patency), and (3) conal septal hypoplasia (as opposed to being well formed). Thus the greater the infundibuloarterial levorotation, the greater the probability that the VSD will be subaortic, that there will be pulmonary outflow tract stenosis, and that the conal septum will be hypoplastic, as in case 26.

Surgical considerations. Surgical management of cases of TGA {S,D,L} should be guided by the clinical status of the patient, which depends on the presence or absence of pulmonary stenosis, and by a comprehensive preoperative and peroperative anatomic analysis.

TGA {S,D,L} with subpulmonary stenosis. Surgical treatment of forms with pulmonary protection is a two-stage repair, including first a modified Blalock-Taussig shunt early in infancy. Complete repair is generally undertaken after age 1 year with a twofold objective: intraventricular rerouting from the left ventricle to the aorta and reconstruction of the right ventricular outflow tract.

Surgical difficulties in rerouting the left ventricular outflow tract are closely related to the size of the VSD. Usually the VSD is large, conoventricular in type, and the construction of a tunnel with a prosthetic patch to establish continuity between the left ventricle and the aorta is easy to do. If the VSD is restrictive, extensive resection of the conal septum is mandatory.^{19, 20}

The possibility of abnormal chordal attachments of the tricuspid valve into the conal septum does not constitute a contraindication to the REV procedure, but requires backward mobilization of the conal septum.²⁰

The reconstruction of the right ventricular outflow tract can be achieved by two techniques: (1) the

classic Rastelli procedure or (2) the direct relocation of the pulmonary artery on the right ventricle, in most of the cases after a Lecompte maneuver.²¹ The relocated pulmonary artery is able to grow, which avoids further reintervention,²² and thus is used in preference to a Rastelli type of intervention. However, the levoposition of the aorta results in an unusual location of the infundibulotomy, which is more rightward than usual and immediately behind the sternum, and this abnormal location entails a risk of growth-related obstruction of the right ventricular outflow tract. This type of complication can also be observed when a conduit is used; this conduit has to be put on the right side of the ascending aorta, in the inner aorticocaval space.

The choice between these two techniques also depends on the possibility of dissecting and freeing the pulmonary arteries. A bilateral Blalock-Taussig shunt precludes a Lecompte maneuver, which may be required for the REV technique. When a REV procedure is anticipated, bilateral Blalock-Taussig shunts should be avoided.

TGA {S,D,L} without subpulmonary stenosis. Surgical management of TGA {S,D,L} with no pulmonary stenosis consists of an anatomic repair, as in TGA {S,D,D}. Few cases of successfully repaired TGA {S,D,L} were reported before the "switch era."²³⁻²⁵ However, the particular anatomic characteristics of TGA {S,D,L} do not prevent performance of the arterial switch operation. The particular disposition of the coronary arteries, as a result of the levoposition of the aortic valve, did not increase the difficulties in coronary ostial relocation in any patient. "Single" right coronary artery does not preclude performance of the arterial switch procedure, but this anomaly has been reported to have a higher postoperative mortality.¹⁰

In conclusion, the anatomic study and surgical results both indicate that TGA {S,D,L}, for the first time delineated as a complex, deserves special attention among the various forms of TGA with atrioventricular concordance.

REFERENCES

1. Van Praagh R, Ongley PA, Swan HJC. Anatomic types of single or common ventricle in man: morphologic and geometric aspects of sixty autopsied cases. *Am J Cardiol* 1964;13:367-86.
2. Van Praagh R, Van Praagh S, Vlad P, Keith JD. Anatomic types of congenital dextrocardia: diagnostic and embryologic implications. *Am J Cardiol* 1964;13: 510-31.

3. Geva T, Van Praagh S, Sanders SP, Mayer JE, Van Praagh R. Staddling mitral valve with hypoplastic right ventricle, crisscross atrioventricular relations, double outlet right ventricle and dextrocardia: morphologic, diagnostic and surgical considerations. *J Am Cardiol* 1991;17:1603-12.
4. Shinpo H, Van Praagh S, Parness I, Sanders S, Molthan M, Castaneda A. Mitral atresia with a large left ventricle and an underdeveloped or absent right ventricular sinus: clinical profile, anatomic data and surgical considerations. *J Am Coll Cardiol* 1992;19:1561-76.
5. Van Praagh R, Van Praagh S. Anatomically corrected transposition of the great arteries. *Br Heart J* 1967;29:112-9.
6. Van Praagh R, Weinberg PM, Srebro JP. Double-outlet left ventricle. In: Moss AJ, Adams FH, Emmanouilides GC, eds. *Heart disease in infants, children, and adolescents*. 4th ed. Baltimore: Williams & Wilkins, 1989:461-85.
7. Serraf A, Lacour-Gayet F, Bruniaux J, et al. Anatomic correction of transposition of the great arteries in neonates. *J Am Coll Cardiol* 1993;22:193-200.
8. Hoyer MH, Zuberbuhler JR, Anderson RH, del Nido P. Morphology of ventricular septal defects in complete transposition: surgical implications. *J THORAC CARDIOVASC SURG* 1992;104:1203-11.
9. DiDonato RM, Wernovsky G, Walsh EP, et al. Results of the arterial switch operation for transposition of the great arteries with ventricular septal defect: surgical considerations and midterm follow-up data. *Circulation* 1989;80:1689-705.
10. Mayer JE, Sanders SP, Jonas RA, Castaneda AR, Wernovsky G. Coronary artery pattern and outcome of arterial switch operation for transposition of the great arteries. *Circulation* 1990;82(Suppl)IV139-45.
11. Melhuish BPP, Van Praagh R. Juxtaposition of the atrial appendages: a sign of severe cyanotic congenital heart disease. *Br Heart J* 1968;30:269-84.
12. Allwork SP, Urban AE, Anderson RH. Left juxtaposition of the auricles with L-position of the aorta: report of 6 cases. *Br Heart J* 1977;39:299-308.
13. Keith A. The Hunterian lectures on malformations of the heart. *Lancet* 1909:433-5.
14. Van Praagh R, Layton WM, Van Praagh S. The morphogenesis of normal and abnormal relations between the great arteries and the ventricles: pathologic and experimental data. In: Van Praagh R, Takao A, eds. *Etiology and morphogenesis of congenital heart disease*. Mt. Kisco, New York: Futura, 1980:271-316.
15. Pexieder T, Pfizenmaier Rousseil M, Prados-Frutos JC. In: Vogel M, Buhlmeyer K, eds. *Transposition of the great arteries 25 years after Rashkind balloon septostomy*. Darmstadt, Germany: Steinkopff, 1992:11-27.
16. Van Praagh S, LaCorte M, Fellows KE, et al. Supero-inferior ventricles: anatomic and angiocardigraphic findings in ten postmortem cases. In: Van Praagh R, Takao A, eds. *Etiology and morphogenesis of congenital heart disease*. Mt. Kisco, New York: Futura, 1980:317-78.
17. Van Praagh R, Perez-Trevino C, Reynolds JL, et al. Double outlet right ventricle {S,D,L} with subaortic ventricular septal defect and pulmonary stenosis: report of 6 cases. *Am J Cardiol* 1975;35:42-53.
18. Van Praagh R. Transposition of the great arteries: history, pathologic anatomy, embryology, etiology, and surgical considerations. *Card Surg State of the Art Reviews* 1991;5:7-82.
19. Kawashima Y, Fujita T, Miyamoto T, Manabe H. Intraventricular rerouting of blood for the correction of Taussig-Bing malformation. *J THORAC CARDIOVASC SURG* 1971;62:825-9.
20. Lecompte Y. Réparation à l'étage ventriculaire: the REV procedure, technique and clinical results. *Cardiol Young* 1991;1:63-70.
21. Lecompte Y, Neveux JY, Leca F, et al. Reconstruction of the pulmonary outflow tract without prosthetic conduit. *J THORAC CARDIOVASC SURG* 1982;84:727-33.
22. Vouhé PR, Tamisier D, Leca F, Ouaknine R, Vernant F, Neveux JY. Transposition of the great arteries, ventricular septal defect, and pulmonary outflow tract obstruction: Rastelli or Lecompte procedure? *J THORAC CARDIOVASC SURG* 1992;103:428-36.
23. Lincoln C, Hasse J, Anderson RH, Shinebourne E. Surgical correction in complete levotransposition of the great arteries with an unusual subaortic ventricular septal defect. *Am J Cardiol* 1976;38:344-51.
24. Matsuda H, Woong Kang J, Kido T, et al. Successful Rastelli operation in infant with complete transposition of great arteries, levoposition of aorta, and subaortic ventricular septal defect. *Ann Thorac Surg* 1982;34:590-3.
25. Quero Jimenez M, Perez Martinez V. Uncommon conal pathology in complete dextrotransposition of the great arteries with ventricular septal defect. *Chest* 1974;66:411-7.